An Interesting Etiology in Childhood Central Diabetes Insipidus HIBERNOMA

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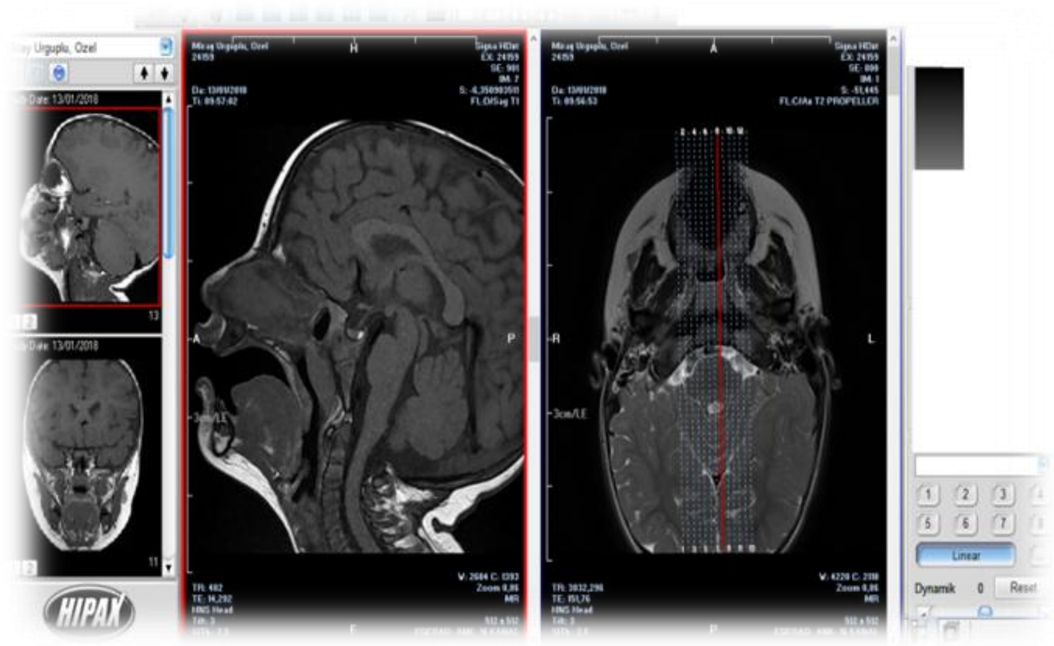
INTRODUCTION

Central Diabetes Insipitus (CDI) results from the inability to secrete ADH secreted by the neurohypophysis system to control water-electrolyte metabolism. ***In the etiology of CDI in childhood, many congenital and acquired CNS tumors (germinoma, pinealoma, craniopharyngioma, optic glioma, AML), infiltrative diseases (langerhans cell histiocytosis, sarcoidosis), infections (meningitis, tbc, encephalitis), autoimmune events, head trauma, idiopathic) can be responsible.*** Mortality and morbidity can be prevented with a timely diagnosis. In this article, a case with CDI due to intracranial occurence which is very rare in etiology is presented and the approach and follow up are discussed.

CASE

Twenty-month-old girl presented with excessive thirst and water intake and frequent urination for about three or four months. The general condition of the patient was good., there were normal physical examination findings. and the somatic development was compatible with age. (Table1). It was learnt that there were no traits in the medical history other than head trauma that had occured twice. While there were no pathological finding in the biochemical parameters, in the follow-up period, the urine osmolarity was found to be polyuric (11.68 ml/kg/h) and the ratio of urine osmolarity to serum osmolarity was found to be 0.27. (Table2). In patients with serum ADH level of <0,5 pmol / l, a 50% increase in urine osmolarity was observed after desmopressin administration. In the non-contrasted and contrast-enhanced pituitary MRI, protein and lipid-containing semisolid structures in the size of 2.5x2 mm were noted in the nasopharyngeal localization, suggesting that this formation led to narrowing of the neurohypophysis volume. (Figure1) This appearance was evaluated as likely to be choristoma-hibernoma.

Central diabetes insipitus was diagnosed in the light of these data.



Desmopressin treatment was begun. Biochemical and imaging analysis were performed.

Table 1: Anthropometric evaluation of the case

AGE	20 months
SEX	FEMALE
PUBERTY	P1
WEIGHT(kg)	10,34(25-50P)
HEİGHT(cm)	86,5(75-90P)
BMI	13,77(0,59)
DIAGNOSIS	CDI*

*CDI; Central Diabetes Insipitus

Table 2: Biochemical and Hormonal Properties

	CASE
ETIOLOGY	*Hibernoma*
COMPLAINT	excessive thirst and water intake and frequent urination
Urine Density (SGU)	*1004*
Polyuria Limit (cc/day)	900
Urine Output (cc/day)	2900
Serum Osmolality (mosm/l)	275
Urine Osmolality (mosm/l)	75
U osm/serum osm	0,27
SerumADH	<0,5
After Desmopressin Test Uosm/serum Osm (2.hour)	0,95
After Desmopressin Test Uosm increase(%)	>%50

DISCUSSION

Hibernoma is a benign soft-tissue well-limited tumor that develops from brownish fatty tissue that is extremely rare in childhood and is observed mostly in adults. ***Up to now, approximately 100 cases have been reported in the literature and 10% are located in the head and neck region.*** Although some publications report that malignant hibernomas may be present, metastasis has not been observed. Although they are benign total excision is suggested when the tumor reaches a certain size due to the possibility of reaching very large dimensions and pressurizing the surrounding tissues. This lesion, which is detected in a very small size in the nasopharynx region of our case, by narrowing the neurohypophysis and causing central diabetes insiput is very important in terms of it being the first in literature. On the other hand, our case is valuable in that it shows the value of imaging in the approach of diagnosis and treatment of childhood diabetes insiputus We also think that the follow-up of this patient in terms of mass size is a necessity because it may have cautionary value in terms of other hormone defects that can develop.